

Congenital heart disease in Nigeria

Necropsy study of 47 cases

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The present study of 47 necropsy cases of congenital heart disease has revealed that cardiac defects occur as frequently in the African as in the non-African. The types of congenital heart disease and the associated non-cardiac malformations are similar to what has been described in non-Africans. Tetrad of Fallot was the commonest type of multiple defect and ventricular septal defect the most common isolated defect in the present series. Two rare types of defects (an accessory third ventricle and congenital tricuspid incompetence) were encountered. The aetiological factors in congenital heart disease in the African environment require further studies.

There has been little published work on the incidence and types of congenital heart defects in Africans. It appears from a few published reports that congenital heart disease occurs in Africans as frequently as in Caucasians. Schrire (1963) in South Africa; Caddell and Connor (1966), Wood, Serumaga, and Lewis (1969) in Uganda; and Caddell and Morton (1967) in Nigeria have reported that the more common types of congenital cardiac defects do occur in the South African Bantu, East and West Africans, respectively. Gupta and Antia (1967), in a study of congenital heart disease in Ibadan, Nigeria, found an incidence of these defects to be 3.5 per 1,000 of all births. Though the total number of subjects studied then was small, the incidence was comparable to those published by European authors (MacMahon, McKeown, and Record, 1953; Gardiner and Keith, 1951; Hay, 1966).

The purpose of the present study is to describe the various types of congenital heart disease seen at necropsy in Nigeria in a period of four and half years (January 1965-July 1969).

Subjects and methods

The necropsy records of the Department of Pathology at the University College Hospital, Ibadan, were examined and all congenital cardiovascular abnormalities for the period abstracted. All the hearts were available for study and the findings were confirmed. The cause of death and associated non-cardiac congenital abnormalities in each

case were also noted. Clinical data obtained from the case notes included age at the time of death, sex, tribe, clinical features, and haemoglobin genotype. A few cases listed in the necropsy register as atrial septal defect or persistent ductus arteriosus were excluded on the grounds that these were either probe-patent foramen ovale, or persistent ductus arteriosus in perinatal deaths unassociated with other cardiac abnormalities.

Results

The total number of necropsies carried out during the period of study was 2,878. The necropsy rate during the period was about 70 per cent. Of the total number of necropsies, 47 cases of congenital heart defects were found, representing 20.8 per 1,000 necropsies. The necropsy incidence of these defects in other series has been reported as 12.9 and 54 per 1,000 in the United States (Leech, 1935; Gibson and Clifton, 1938), 21.1 and 6.4 per 1,000 in Singapore and Uganda respectively (Muir, 1960; Wood *et al.*, 1969). All the subjects in this study were Nigerians. Of the 47 patients, 44 were Yorubas, and of the remaining 3, 2 were Ibos from the Eastern and 1 was a Bini from the Mid-Western Nigeria. The University College Hospital is situated in the Western State of Nigeria and is therefore easily accessible to its inhabitants who are Yoruba ethnic group.

The various types of defects, their distribution, the sex incidence, and the number of associated non-cardiac congenital malformations are summarized in Table 1. Tetrad of Fallot was the most common defect (21% in

TABLE 1 *Types of congenital cardiac defect, sex, and associated non-cardiac malformations*

Type of defect	No. of cases	% of total	Sex		No. with associated non-cardiac malformations
			M	F	
A					
<i>Isolated defect</i>					
Ventricular septal defect	8	17.0	3	5	4
Atrial septal defect	4	8.5	1	3	
Persistent ductus arteriosus	5	10.6	3	2	2
Pulmonary stenosis with intact ventricular septum	3	6.4		3	1
Tricuspid valve lesion:					
a) Ebstein's malformation of tricuspid valve	1	2.1		1	
b) Congenital tricuspid incompetence	1	2.1			
B					
<i>Multiple defects</i>					
Tetrad of Fallot	10	21.3	6	4	3
Tricuspid atresia	3	6.4	2	1	2
Mitral atresia	2	4.3	1	1	
Transposition of great arteries	3	6.4	3		2
Single ventricle	2	4.3	2		1
Aortic anomalies	2	4.3	2		1
Persistent left superior vena cava draining into coronary sinus + ventricular septal defect and persistent ductus arteriosus*	1	2.1	—	—	1
Persistent truncus arteriosus plus atrial septal defect	1	2.1		1	
Origin of aorta from right ventricle + ventricular septal defect + accessory third ventricle	1	2.1		1	
Total	47	100	22	24	17

* Intersex.

the entire series. Of the isolated defects, ventricular septal defect was the commonest and persistent ductus arteriosus the next.

There was no significant difference in the overall sex incidence (22 male and 24 female).

The sex of a 1-day old infant was indeterminate.

Associated non-cardiac congenital malformations occurred in 17 (36%) patients (Table 2). Seven malformations (41%) which oc-

TABLE 2 *Types of congenital cardiac defects and associated non-cardiac congenital anomalies*

Type of cardiac defect	Associated non-cardiac anomalies
Ventricular septal defect	Imperforate anus, bifid uterus, gall-bladder agenesis
Ventricular septal defect	Microcephaly, odd facies, and high arched palate
Ventricular septal defect	Down's syndrome
Ventricular septal defect	Tracheo-oesophageal fistula, duodenal atresia
Persistent ductus arteriosus	Duodenal atresia
Persistent ductus arteriosus	Hydrocephalus, bilateral talipes
Pulmonary artery stenosis with intact septum	Occipital encephalocele, polycystic kidneys
Tetrad of Fallot	Inguinal hernia
Tetrad of Fallot	Inguinal hernia
Tetrad of Fallot	Tracheo-oesophageal fistula
Aortic hypoplasia + ventricular septal defect + persistent ductus arteriosus + anom. venous return	Right-sided stomach, spleen, and liver
Single ventricle	Bladder neck obstruction due to valves
Complete transposition of great arteries	Exomphalos
Complete transposition of great arteries	Tracheo-oesophageal fistula
Tricuspid atresia	Arachnodactyly, high arched palate
Tricuspid atresia	? Marfan's syndrome
Tricuspid atresia	Renal cysts
Persistent superior vena cava + ventricular septal defect + persistent ductus arteriosus + bicuspid pulmonary valve	Cleft lip and palate, absent eyeballs, bilateral syndactyly

TABLE 3 Age when death occurred in 47 cases of congenital heart disease

Type of defect	Week		Month			Year				Total no. of cases
	0-1	2-4	2-3	4-6	7-11	1-3	4-6	7-9	10-	
Ventricular septal defect	2		2	2	1	1				8
Atrial septal defect		1	1				1		1	4
Persistent ductus arteriosus	2	2	1							5
Pulmonary stenosis with intact ventricular septum	2					1				3
Ebstein's defect of tricuspid valve						1				1
Congenital tricuspid incompetence							1			1
Tetrad of Fallot	1			1	1	2	3	1	1	10
Tricuspid atresia		1		1				1		3
Persistent truncus arteriosus	1									1
Single ventricle	1	1								2
Transposition of great arteries	1	1				1				3
Aortic anomalies	1	1								2
Origin of aorta from right ventricle + ventricular septal defect + accessory third ventricle							1			1
Mitral valve atresia	1		1							2
Persistent left superior vena cava + ventricular septal defect + persistent ductus arteriosus	1									1
Total	13	7	5	4	2	6	6	2	2	47

occurred in the alimentary tract included cleft lip and palate (1), tracheo-oesophageal fistula (3), duodenal atresia (1), imperforate anus (1), and right-sided stomach (1). None of the malformations occurred with any significant frequency in the different types of cardiac defects.

The age at which death occurred in the 47 patients is listed in Table 3. The majority of these children, 31 (66%), died in the first

year of life. Death in the neonatal period occurred in 20 patients (43%). Most of the older patients died in the third to sixth year of life. The causes of death in 41 patients are presented in Table 4. It is evident that bronchopneumonia and postoperative complications were the commonest causes of death. Of the 9 patients who died after surgical operation, 5 had serious associated malformations for which an operation had to be performed

TABLE 4 Causes of death in 41 cases of congenital cardiac defect

Type of defect	No. of cases	Causes of death						
		Broncho-pneumonia	Subacute bacterial endocarditis	Cerebral abscess	Heart failure	Still-birth	Post-operative	Others
Ventricular septal defect	8	4					1	3
Atrial septal defect	4	2			1			1
Persistent ductus arteriosus	5	2			1		1	1
Pulmonary stenosis with intact ventricular septum	3	1				1		1
Tetrad of Fallot	10	1	1	1			5	2
Ebstein's malformation of tricuspid valve	1				1			
Tricuspid atresia	3			1				2
Mitral atresia	2	1			1			
Single ventricle	2			1				1
Complete transposition of great vessels	3	1					2	
Total	41	12	1	3	4	1	9	11

in the neonatal period. In these 5 patients the immediate cause of death was not the cardiac defect. The remaining 4 patients had tetrad of Fallot with severe cyanosis and conspicuous decrease in exercise tolerance. They all died soon after various palliative surgical procedures were carried out to increase the pulmonary circulation. All 3 patients who died from cerebral abscess had the cyanotic type of cardiac defect. Subacute bacterial endocarditis occurred in one patient with tetrad of Fallot. Eleven patients died from various causes which included gastroenteritis, neonatal tetanus, purulent bacterial meningitis, and kwashiorkor.

The haemoglobin genotype was determined in 21 subjects and the distribution was 12 AA, 7 AS, and 2 SC. No one type of cardiac defect was associated with these genotypes in significant frequency.

The isolated congenital cardiac defects (Table 1) consisting of ventricular septal defect, atrial septal defect, persistent ductus arteriosus, pulmonary stenosis with intact ventricular septum, and Ebstein's malformation of the tricuspid valve were of the usual type. The single case of congenital tricuspid incompetence with unusual clinical and pathological features has already been reported (Antia and Osunkoya, 1969).

Of the multiple cardiac defects, tetrad of Fallot was the commonest type. In addition to the recognized combination of the defects in tetrad, 2 subjects with much less cyanosis had persistent ductus arteriosus as well.

There were two cases of aortic anomalies. One had aortic hypoplasia, ventricular septal defect, persistent ductus arteriosus, and anomalous venous drainage into the right atrium, while the other had a ventricular septal defect, complete interruption of the aortic arch, and bicuspid aortic valve. Both patients died in the neonatal period.

Three patients had tricuspid atresia as the dominant defect. The associated cardiac defects in one were a persistent ductus arteriosus, atrial septal defect, and hypoplasia of the pulmonary artery; in another, atrial septal defect, ventricular septal defect, and hypoplasia of the pulmonary artery; in the third, pulmonary stenosis, a tiny ventricular septal defect, and a large left ventricular cavity.

Mitral valve atresia was associated with a single (right) ventricle, large atrial septal defect, and persistent ductus arteriosus in one subject, and in the other, an atrial septal defect, and the aorta and pulmonary artery arising from a single ventricle which, anatomically, was a right ventricle.

There were two cases with a single ven-

tricle. One patient had hypoplasia of the aorta and a persistent ductus arteriosus and another had a hypoplastic pulmonary artery.

Of the 3 patients with transposition of the great arteries, one had multiple muscular ventricular septal defects and bicuspid aortic valve; the second had a ventricular septal defect and pulmonary stenosis; and the third had a ventricular septal defect, persistent ductus arteriosus, and infundibular pulmonary stenosis.

The single case with persistent truncus arteriosus was Edward's type II with atrial septal defect.

Other cardiac defects in the single patient with persistent left superior vena cava draining into the coronary sinus included a persistent ductus arteriosus, ventricular septal defect, and bicuspid pulmonary valve. This patient died at the age of 6 days. The sex was indeterminate.

One patient had an unusual complex defect consisting of the origin of the aorta from the right ventricle, an accessory third ventricle from which a hypoplastic pulmonary artery arose, and a tiny ventricular septal defect. This case will be reported in detail elsewhere.

Discussion

The present study, which is part of an effort to determine as comprehensively as possible the types of congenital heart disease in the African, clearly shows that most of the defects seen elsewhere occur in Nigerians. The incidence of the different types of cardiac defects appears to be the same as in Caucasians; however, no conclusion on this aspect of the disease can be made from the relatively small number of cases in the present study.

The incidence of non-cardiac malformations associated with congenital heart disease has been reported by several authors. Abbott (1927) found associated anomalies in 18.8 per cent of her 1,000 cases; Gibson and Clifton (1938) found an incidence of 27 per cent; Wood *et al.* (1969) found an incidence of 20 per cent. In the series reported by MacMahon *et al.* (1953) the incidence of associated anomalies was 21 per cent. In the present study the incidence was 36.1 per cent, and the anomalies were similar to those reported by others. There was, however, a strikingly high incidence (31%) of anomalies in the alimentary tract. A similarly high incidence (67.7%) of alimentary tract anomalies was found by MacMahon *et al.* (1953). This high incidence is to be expected, since embryologically the heart and the alimentary tract are formed about the same time and any injury to one

would be expected to affect the other. The aetiological factors in this environment are not known and require further studies.

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